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Mesenchymal chondrosarcoma of the mandible: A case report

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ABSTRACT

Mesenchymal chondrosarcoma as an aggressive type of chondrosarcoma shows a characteristic biphasic histopathologic pattern. The head and neck region is included a high proportion of extra skeletal sites. Very rare examples of Mesenchymal Chondrosarcoma involving the mandible have been described. Based on fragmented or tiny specimens, the diagnosis of this lesion has been remained a challenge because the specimens may contain only one of the two neoplastic elements. We report a rare case of mesenchymal chondrosarcoma of the mandible in a 19 years old male with delaying in diagnosis due to massive extension of the tumor to the soft tissues.

Keywords: Mesenchymal chondrosarcoma; Mandible; Neoplasm.

Introduction

Mesenchymal chondrosarcoma (MC) as a rare subtype of chondrosarcoma was first described in 1954 by Lichtenstein and Bernstein [1]. It represents around 1% of all chondrosarcomas. It usually occurs in second and third decade of life. Females are more frequently affected than males [1,2]. Facial bones particularly the jaws (22%-27%) and ribs appear to be involved more often and the maxillary sinus is the most commonly involved sites. Swelling and pain, often of fairly short duration, are the most common symptoms [1,2]. Other symptoms are including nasal obstruction and paresthesia [3]. This neoplasm presents both local aggressive behavior and a high metastatic potential. The long term prognosis is extremely poor. Radiographically, the tumor commonly represents as an ill-defined radiolucency with or without stippled calcification. However, some cases may manifest predominantly radiopaque, especially within the maxilla [3]. Characteristic histological features of an established MC include sheets or clusters of highly undifferentiated, small, ovoid cells that alternate with small zones of neoplastic cartilage [4]. We report a mandibular MC in 19-year-old male with postponing the treatment due to delay in diagnosis.

Case Report

A 19 years old male was referred to Department of Oral and Maxillofacial Pathology, with complaint of painful welling from last four months in anterior region of the lower jaw. He noticed a history of antibiotic consumption by self-prescriptive for decreasing the pain about two months ago. Also he had a history of parotid malignancy and associated radiotherapy. In extra oral examination the patient showed facial asymmetry due to a large swelling of anterior of mandibular jaw extending from midline to left body of the mandible, with normal color and intact overlying skin (Fig 1a). On palpation, it was tender, non-compressible non-pulsatile and firm in consistency. Intra-oral examination showed an ulcerated mass extending from left lower second premolar to midline involving labial vestibules. Mucosa over mass was reddish in color in non-ulcerated areas (Fig 1b).

Mobility and displacement of teeth were not seen. Periapical view of anterior mandibular teeth revealed PDL widening in association with radiolucency of nearby teeth (Fig 2a). Furthermore, on panoramic view, an ill-defined lesion with was evident (Fig 2b). Incisional biopsy of the lesion was done. Microscopic examination showed a tumoral tissue composed of admixture of sheets of undifferentiated cells demonstrate in ground to oval cells with hyperchromatic nuclei and few amount of cytoplasm are arranged in lobular pattern and foci of well differentiated cartilaginous tissue. Foci of hemangiopericytoma-like pattern around sinusoidal vascular channels that lined by a single layer of endothelium are also evident (Fig 3). Foci of well differentiated, benign-shaped cartilaginous tissue, with central ossification were present (Fig 3). Although these features were in favor of MC, for definitive diagnosis Immunohistochemical studies were mandatory. Immunohistochemical findings showed positive reaction for vimentin and negative for CD99 and Ewing sarcoma was confirmed (Fig 3). Based on IHC results and also correlation of the clinical, radiological and the histopathological features the diagnosis of MC has been confirmed. The patient undergone the partial mandibulectomy and post-operative radiotherapy but unfortunately died after 14 months after surgery due to several distant metastasis.

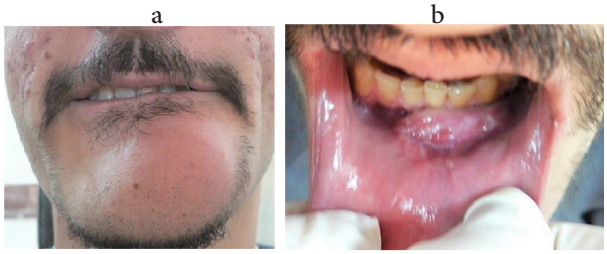

Figure 1. Pre-operative photograph of the patient. a) Extra oral view shows mandibular swelling due to facial asymmetry. b) Intra oral view shows an exophytic lesion in anterior buccal vestibule of mandible.

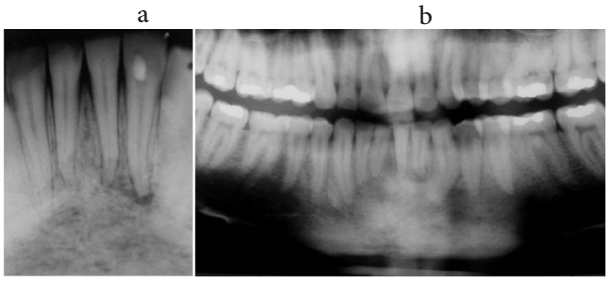

Figure 2. Radiographic views of the lesion. a) Periapical image. Note PDL widening of the teeth. b) Panoramic image shows large mixed lesion with ill-defined borders.

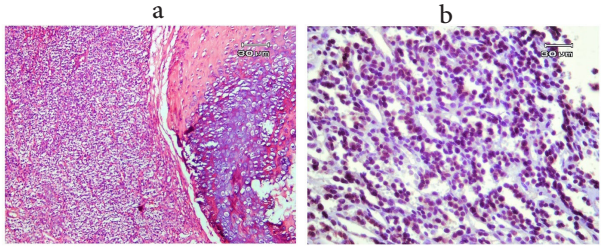

Figure 3. a) Microscopic examinations of the lesion show sheet of round cells and cartilage tissue with central ossification. b) Immunohistochemistry study was negative for CD99.

Discussion

Mesenchymal chondrosarcoma is a malignant tumor [5]. We reported a case of mandibular MC in a 19 years old man with high aggressiveness potential and very short survival rate. The staining characteristics of the cartilaginous areas are indistinguishable from those of other forms of chondrosarcoma [4]. Although characteristic histopathologic features of MC presents no particular diagnostic problem, recognition of this tumor may be difficult with small biopsy or needle biopsy specimens that shows only one of the two tissue entities [6]. In particular, tumors without the cartilaginous element may be misdiagnosed for ES/PNET, malignant hemangiopericytoma, or poorly differentiated synovial sarcoma with a hemangiopericytoma-like pattern. Al-

though it has been reported well differentiated cartilage in one case of benign hemangiopericytoma, it is a rare finding; and as a rule the presence of cartilage excludes the diagnosis of hemangiopericytoma [7,8,9]. Metaplastic cartilage may also occur in poorly differentiated synovial sarcoma, but it is much less common than foci of calcification or bone. Careful search for a biphasic pattern or epithelial differentiation with antibodies against cytokeratin or epithelial membrane antigen is necessary in difficult cases [7,8 and 9].

Immunohistochemically, the cartilaginous portion of the tumor typically shows strong S-100 protein positivity, whereas only isolated cells in the undifferentiated areas stain for this antigen [10]. Moreover, as in other round cell tumors, the undifferentiated cells may stain for neuron-specific enolase and Leu-7. Stains for desmin, actin, cytokeratin, and epithelial membrane antigen, synaptophysin, CD31 and CD34 are typically negative [11,12 and 13]. In the absence of the cartilaginous foci, the undifferentiated areas may closely resemble other round cell sarcomas, particularly ES/PNETs. Although early reports on the product of the MlC2 gene (CD99) indicated that the undifferentiated areas of extra skeletal mesenchymal chondrosarcoma were negative for this antigen [13,14]. Bret et al showed MC has phenotypic features corresponding to the early condensational phase of cartilaginous differentiation. More important, Sox 9 may serve as a useful tool in the differentiation of small cell malignancies [15].

Lee et al showed in contrast to Ewing sarcoma, small cell osteosarcoma and mesenchymal chondrosarcoma lack FLI-1 immunoreactivity. FLI-1 is therefore useful in the differential diagnosis of small round blue cell tumors of the bone [16]. The most approach for treatment is wide surgical excision along with chemotherapy and radiotherapy [17]. According to Nakashima et al., extensive resection has less recurrence and a better survival rate than limited surgical resection [18]. Prognosis of MC is poor because the lesion has a high tendency for late recurrence either locally or metastasizes [19]. Time and accuracy are very important in diagnosing of this lesion. The accuracy of the diagnosis depends on histopathological examination of the multiple sections along with IHC. Recurrence and metastasis are very frequent with MC, however follow-up for a long time should be done. In our case, mandibular MC with late diagnosis has led to a poor prognosis because of the aggressive nature of the lesion. It means that the more accurate and early diagnosis of MC is very important to have a better prognosis for the patients.

Conflict of Interest

There is no conflict of interest to declare.

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